Factors Associated with Delay in Diagnosis of Patients with Fibromuscular Dysplasia: A Report From the United States Registry for Fibromuscular Dysplasia

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Background: Fibromuscular dysplasia (FMD) is an uncommon disease of medium-sized arteries that may result in stenosis, dissection or aneurysm. It is unclear what factors affect delay between first signs/symptoms and diagnosis.

Methods: Patients enrolled in the FMD registry from 10 U.S. sites were stratified into three groups based upon the length of time between first sign/symptom and diagnosis.

Results: Of the 615 total patients enrolled in the FMD registry, 538 patients had sufficient data for analysis. The mean length of time from first reported clinical sign/symptom to diagnosis was 3.6 ± 7.4 years. As outlined in the table below, FMD patients with greater delay in diagnosis were younger at first sign/symptom and older by the time of diagnosis. Patients with a greater delay in diagnosis were more likely to present with hypertension, had earlier onset and greater family history of hypertension. Furthermore, they had a greater mean number of blood pressure medications and were more likely to take an ARB, diuretic or alpha blocker when compared to patients with a smaller gap between first sign/symptom and diagnosis. Conversely, FMD patients with a shorter time to diagnosis were more likely to have presented with a carotid or renal artery dissection.

	Time between first sign/symptom and diagnosis			
	< 3 years N (%)	3 – 5 years N (%)	> 5 years N (%)	p-value
N	383/538 (71.2)	55/538 (10.2)	100/538 (18.6)	
Age at first sign/symptom (mean ± SD)	50.4±13.3	45.7±12.4	39.1±15.8	<0.0001
Age at diagnosis (mean ± SD)	50.6±13.3	49.5±12.4	55.3±13.6	0.0041
Family history of hypertension	241/333 (72.4)	41/53 (77.4)	82/94 (87.2)	0.0087
Age at onset of hypertension (mean ± SD)	44.8±13.9	39.6±13.6	38.9±16.0	0.0056
Presenting Symptoms or signs				
Headache	195/339 (57.5)	31/53 (58.5)	54/94 (57.4)	1.0
Hypertension	229/360 (63.6)	34/54 (63.0)	75/96 (78.1)	0.021
Pulsatile tinnitus	106/321 (33.0)	20/50 (40.0)	31/88 (35.2)	0.60
Hemispheric TIA	35/330 (10.6)	4/51 (7.8)	7/93 (7.5)	0.67
Stroke	30/336 (8.9)	0/52 (0)	10/94 (10.6)	0.028
Cervical Bruit	86/311 (27.7)	12/50 (24.0)	18/90 (20.0)	0.34
Carotid artery dissection	64/326 (19.6)	2/50 (4.0)	7/91 (7.7)	0.0008
Renal artery dissection	17/327 (5.2)	1/52 (1.9)	0/92 (0)	0.0350

Conclusion: The delay from first sign/symptom to diagnosis in FMD patients is prolonged and associated with having hypertension. This study suggests that the role of FMD is under-

appreciated in patients who present with early onset hypertension. Patients with an acute arterial dissection are more likely to have a timely diagnosis of FMD. Further effort may be needed to increase physician awareness of FMD to more effectively diagnosis this disease and expedite appropriate treatment.